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# BONY SYNGNATHIA (CONGENITAL FUSION OF MAXILLA AND MANDIBLE)

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***ABSTRACT***

Congenital bony fusion of the jaws (syngnathia) without any other anatomic oral anomalies is a very rare condition. Numerous cases with combination of cleft palate, aglossia, and soft or bony adhesion between the maxilla and mandible have been reported. Syngnathia could also occur with popliteal pterygium syndrome and van der Woude syndrome. This report presents a case of syngnathia who was attend Maxillofacial surgery Department at Ramadi Teaching Hospital, Anbar Province, Iraq, with bilateral maxillo-mandibular inter-alveolar adhesion, with no other intra-oral anomalies.

**KEY WORDS:** Syngnathia, bony fusion, congenital defect, maxilla, mandible.

## INTRODUCTION

Syngnathia is a rare congenital anomaly involving fusion of maxillary and mandibular bones. The fusion may be due to soft tissue adhesions between the two or a true bony fusion between maxilla and mandible[1,2]. The effected new born child is presenting with difficulties in the airway protection and maintenance as well as feeding problems[3].

Most have other associated anomalies like popliteal pterygium syndrome, Vander Woude syndrome requiring concurrent management. Surgical management involves division of the bony fusion or break down of the adhesions in the first few days of life. Depending upon the severity, these children present formidable anaesthetic challenges. The congenital bony fusion of the maxilla and mandible (bony syngnathia), especially as an isolated occurrence, is a very rare condition. Syngnathia mostly appears in association with other anatomic oral and maxillofacial anomalies. About few such cases have been reported in the literature in combination with cleft lip, cleft of hard and soft palate, aglossia, popliteal pterygium syndrome[1], van der Woude syndrome, aglossia-adactylia syndrome[2], oral soft tissue synechiae, hypoplasia of the proximal mandible, clefting of mandible, bifid tongue, hemifacial microsomia, small or absent tongue, temporomandibular (zygomaticomandibular) fusion and some other regional and systemic anomalies[3-8] .

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## CASE PRESENTATION

A newborn male ( weight 2800g) of 3 days of age was referred to the Department of Maxillofacial Surgery of Ramadi Teaching Hospital, Ramadi city, Anbar Province, Iraq; for evaluation of the fusion of maxilla and mandible that prevented oral feeding. Medical consultation and laboratory tests revealed low blood glucose. Urgently thorough clinical and radiographical examinations were done and start feeding via a nasogastric (NG) tube and the patient was maintained on humidified oxygen by mask. There was no other associated local or systemic anomaly. Clinical examination revealed severe trismus due to adhesion of the jaws, which extended bilaterally from deciduous canine area to the molar regions posteriorly and revealed a bony fusion between the maxilla and the mandible involving the entire alveolar margin with a small gap about 18 mm on left side anteriorly in the canine region. The radiographs were obtained with difficulty, revealed bilateral bony fusion of the maxilla and mandible, Very feeble motion was palpable over each temporomandibular joint (TMJ). Family history showed no similar affliction could be elicited in the past generations of either the parents and maternal and paternal history was negative for any facial cleft. All other siblings were normal.( Fig. 1 and 2)

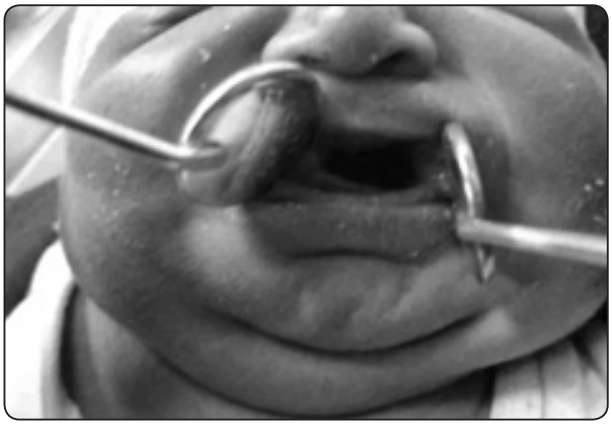

FIG. (1) Bilateral fusion of both jaws

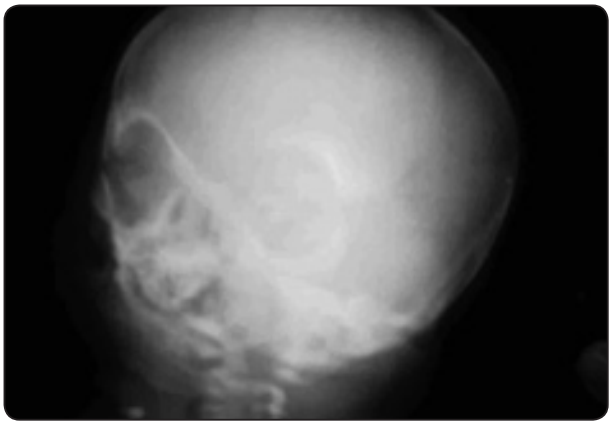

FIG. (2) PA radiograph of the patient

## DISCUSSION

Fusion defects of the maxilla and mandible including other anatomic oral abnormalities are not common. They may be of the connecting tissue either fibrous or bony [1,2] Soft tissue fusion [9,10] (synechiae) have been extensively reviewed by Gartlan et al. (1993) and were classified as buccopharyngeal membrane remnants or as ectopic membranes on the basis of their presumed origin [11] . Bony fusion (syngnathia), particularly its isolated occurrence, is extremely rare. The very few cases reported in the literature are mostly inadequate in description, inconsistent and confusing in nomenclature and with limited useful conventional imaging [4,10]. The cause of congenital bony syngnathia is not certain. In contrast, the review of five cases presented by Dawson et al (1997) and previously reported cases provide no evidence of any familial tendency, history of drug and toxin exposure or consanguinity[3]. Congenital bony syngnathia can be clinically recognized and diagnosed at or after the birth of the affected neonate without any exception [3,7]. The adequate useful conventional radiography and / or CT scan can support the clinical recognition of this condition and its nature, which causes inability to open the jaws. The management of patients with congenital fusion of maxilla and mandible varies according to the nature and extent of the abnormalities.

The condition is problematic and interferes with feeding, breathing, general health of the patient (aspiration pneumonitis), growth and development, induction of anesthesia. The airway is the first priority to be secured in the management of any newborn with trismus. Thereafter, feeding problems should be overcome by placing a nasogastric or gastrotomy tubes. Since the occurrence of bony fusion of the maxilla and mandible is extremely rare, and there is high rate of association between bony syngnathia and other regional and systemic malformations[3,4,6,8,12], the patient should be under the supervision of a team of clinicians skilled in the diagnosis and appropriate treatment of congenital oral and maxillofacial anomalies [3] . Surgical division of the bony fusion, under general anesthesia (blind intubation or via tracheostomy) is the optimal treatment for the simple syngnathia in isolated occurrence or cases with the presence of other anatomic abnormalities[6,8,12] . Proper physical therapy should be commenced immediately and the infant should be encouraged to feed normally as soon as possible[3]. Maxillomandibular fusion is a rare group of anomalies varying in severity from simple mocusal adhesions (synechiae) to extensive bony fusion (syngnathia). Proper physical therapy and feeding should be resumed as soon as possible after the surgery. The significant points about the case reported were; it was an isolated pure bony fusion without any associated local (cleft lip/palate) or systemic anomalies; only few cases exist in the world literature so far.

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